corrected vision regularly in each eye. If one eye is two or more lines worse than the other, with no other apparent explanation, it is probably amblyopic and the child needs occlusion treatment of the preferred eye. The risk of amblyopia is greatest during the first year of life and declines rapidly after the age of five.

Providing optimum care is provided, the visual prognosis is good. In Kenya, 47% of eyes achieved 6/18 or better and only 5% were less than 6/60.[1] Almost all these children will be able to attend a normal school (Fig.1).

Complications

Every child who does not have a posterior capsulotomy will develop posterior capsule opacification. This can be treated by making an opening in the capsule with a Nd:YAG laser or a needle. Alternatively, the posterior capsule and anterior vitreous can be removed with a vitrector. If the capsule is opened without removing the vitreous, the opacification may recur on the anterior hyaloid face. Loss of vision in one eye from increasing capsule opacity will be asymptomatic and the only way to detect this is by regular examinations.

Glaucoma may occur after lensectomy, particularly if it is carried out in the first week of life. This glaucoma is very difficult to treat and frequently leads to blindness. Delaying surgery until after the child is 3–4 months old makes it unlikely that the eyes will recover 6/6 vision but it reduces the risk of glaucoma.

Retinal detachment is more common in eyes that have had surgery for congenital cataract. It often occurs very late, on average 35 years after the operation. If any patient complains of sudden loss of vision, even if it is years after their operation for congenital cataract, it should be assumed to be due to retinal detachment until proven otherwise.

Conclusion

The management of congenital cataract is complex, and should only be carried out in specialist centres. However, every eye worker can play a role by assisting with case finding and follow-up.

Reference

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Evidence for the Effectiveness of Interventions for Congenital, Infantile and Childhood Cataract

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Introduction

Certain groups are often excluded from trials of new interventions, typically pregnant women and children, but also people unable to give informed consent. Children are not included perhaps because of a distaste for 'experimenting' on little ones and a reluctance to admit to clinical uncertainty when faced with anxious parents.

Unfortunately such attitudes lead to continuing uncertainty about the effectiveness of key interventions in these population subgroups who are often, ironically, the subject of our special concern.

Cataract in children is an important cause of childhood blindness and treatment can make a difference if it can be delivered effectively and in time. But there are many questions about how this is best achieved – clinical questions which need good evidence for an answer. And before addressing these, there are others – about how best to detect cataracts in babies (there are no randomised trials as yet) and whether or not there is any potential for prevention. Immunisation for rubella is relevant, and, of course, understanding genetics.

Treatment of Bilateral Cases

There are numerous surgical procedures described for the treatment of cataract including peripheral iridectomy (for central opacities), needling and aspiration, lensectomy, optic captured posterior chamber intraocular lens after phaco-emulsification (termed 'bag in the lens' procedure by one group). So what is the best procedure in terms of visual outcome, short and long term complications and cost-effectiveness? A Cochrane Systematic review was published in 2001 asking the question "What is the effectiveness of surgical interventions for bilateral congenital cataract". As in most Cochrane reviews, only the best evidence was included, i.e. randomised controlled trials. Only one trial was found comparing pars plana lensectomy to lens aspiration and primary capsulotomy. Though both groups did well in terms of visual outcome, there were more complications in the aspiration group but follow-up was not long enough to address the important concern about late glaucoma after lensectomy. The review is now in the process of being updated. Since it was published, clinical practice has been changing and lens implantation in the bag with or without primary posterior capsulotomy or with capture of the optic within anterior and posterior capsulorhexis is becoming more common. The age at which surgeons are happy to intervene is also falling.

The situation is made more complex by the fact that several different parameters are being modified simultaneously so that it is hard to determine which are the key components to improved outcome.

Since 2001, seven new trials have so far been identified including a large one from China comparing acrylic and polymethyl methacrylate lenses (though it is not clear if this was truly randomised) and four on various aspects of technique relating to optic capture. The other two are on the use of trypan blue for capsulorhexis and a comparison of two methods of hydrodissection.

Treatment of Unilateral Cases

The treatment of unilateral congenital cataract is another question and is the subject of much discussion though so far there are no trials. A study examining the feasibility of randomising children in USA to intraocular lens or contact lens correction of aphakia has been published in the Journal of Amerian Association of Pediatric Ophthalmology and Strabismus (AAPOS). This article also describes the considerable amount of stress that such interventions place on both the child and parents when the results of preserving useful sight in the cataractous eye are not great.

The treatment of stimulus deprivation amblyopia in both unilateral and bilateral cases is also in need of good evidence of effectiveness and a title for a Cochrane review on this subject has been registered.

Conclusion

This remains an issue of intense importance in the control of childhood blindness and, as

yet, the quality of evidence is relatively poor. But the signs are encouraging and there are indications that those involved in this work are increasingly aware of the need for it. It is of particular importance in countries with emerging economies where the volume of need is substantial and the opportunities for high quality research now exist.

Evaluation of screening procedures for congenital cataracts

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AIM: To evaluate the efficacy of two different Swedish screening procedures for early detection of congenital cataracts in comparison with no screening. METHODS: Children born between January 1992 and December 1998 in Swedish regions with an established eye-screening routine procedure, diagnosed with congenital cataract, and operated on before 1 year of age, were included in a retrospective study. Age at referral and age at time of the operation were compared between regions using different screening procedures: screening in the maternity wards (Region 1), at the well-baby clinics (Region 2) and one region without any screening (Region 3). RESULTS: Seventy-two children were included in the study. Concerning early diagnosis and surgery, Region 1 differed significantly from Regions 2 and 3, which were more similar and were combined for further analysis. The difference in detected cases was greatest at 21 days of age (55% vs 18%; $p < 0.001$), but persisted even at 100 days of age (78% vs 64%; $p < 0.2$). Region 1 screening resulted in more and earlier cases detected than the other two regions (22 vs 15 per 100,000 births). In 72% of all cases, surgery was performed in response to referrals from either the maternity wards (36%), or the well-baby clinics (36%). However, half of the cases from the well-baby clinics were detected too late, i.e. at > 100 days. CONCLUSION: Eye screening in the maternity ward is preferable to well-baby clinic screening and to no screening at all, since it leads to early detection. Screening should also be performed routinely at well-baby clinics within the period when successful treatment is possible.

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Outcome of lens aspiration and intraocular lens implantation in children aged 5 years and under

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AIMS: To determine the visual outcome and complications of lens aspiration with intraocular lens implantation in children aged 5 years and under. METHODS: The hospital notes of all children aged 5 years and under, who had undergone lens aspiration with intraocular lens implantation between January 1994 and September 1998, and for whom follow up data of at least 1 year were available, were reviewed. RESULTS: Of 50 children who underwent surgery, 45 were eligible based on the follow up criteria. 34 children had bilateral cataracts and, of these, 30 had surgery on both eyes. Cataract was unilateral in 11 cases; thus, 75 eyes of 45 children had surgery. Cataracts were congenital in 28 cases, juvenile in 16, and traumatic in one case. The median age at surgery was 39 months (range 11-70 months). Follow up ranged from 12–64 months (median 36 months). Of 34 children with bilateral disease, 25 (73.5%) had a final best corrected visual acuity of 6/12 or better, while seven (20.5%) achieved 6/18 or less; in one child the vision improved from UCUSUM to CSM but another, who had only one eye operated on, was unable to fix or follow with this eye preoperatively or 2 years postoperatively. Of 11 children with unilateral cataract, five (45.5%) had a final best corrected visual of 6/12 or better, and six (54.5%) 6/18 or less. A mild fibrinous uveitis occurred in 20 (28.2%) eyes in the immediate postoperative period, but resolved with topical steroids. One child had a vitreous wick postoperatively requiring surgical division. Glaucoma, endophthalmitis, or retinal detachment have not been observed so far in any patient postoperatively. CONCLUSION: From this series the authors suggest that, in children aged 5 years and under, lens aspiration with intraocular lens implantation is a safe procedure, with a good visual outcome in the short term. Further studies are needed to investigate these outcomes in the long term.

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Is early surgery for congenital cataract a risk factor for glaucoma?

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AIMS: To estimate the risk of aphakic glaucoma after lensectomy for congenital cataract and its association with surgery within the first month of life. METHOD: A retrospective case notes review was conducted of all patients who had lensectomy for congenital cataract during their first year of life at Great Ormond Street Hospital between 1994 and 1997. Patients with pre-existing glaucoma, anterior segment dysgenesis, and Lowe syndrome were excluded. The risk of aphakic glaucoma after surgery was estimated using Kaplan-Meier survival analysis. RESULTS: 80 patients, undergoing 128 lensectomies were eligible. Of these, six patients (nine eyes) were lost to follow up. Based on eye count, the risk of glaucoma by 5 years after lensectomy was 15.6% (95% CI 10.2 to 23.4). Based on patient count, the 5 year risk of glaucoma in at least one eye following bilateral surgery was 25.1% (95% CI 15.1 to 40.0). The incidence of glaucoma remained at a constant level for the first 5 years after surgery. After early bilateral lensectomy, within the first month of life, the 5 year risk of glaucoma in at least one eye was 50% (95% CI 27.8 to 77.1) compared to 14.9% (95% CI 6.5 to 32.1) with surgery performed later (log rank test, $p = 0.012$).

There was no significant difference (Kolmogorov-Smirnov test: unilateral lensectomy $p = 0.587$, bilateral lensectomy $p = 0.369$) in 5 year visual outcomes between eyes operated before and after 1 month of age. CONCLUSION: Bilateral lensectomy during the first month of life is associated with a higher risk of subsequent glaucoma than with surgery performed later. The reason for this is unclear but it may be prudent, in bilateral cases, to consider delaying surgery until the infant is 4 weeks old. As the incidence of glaucoma is similar for each year after surgery, long term glaucoma surveillance is mandatory.

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